Spontaneous aortic arch thrombosis in a neonate

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Spontaneous neonatal aortic arch thrombosis is extremely rare and has significant mortality and morbidity. We report our recent experience with a newborn diagnosed with this condition. Human cytomegalovirus (HCMV) infection may have played a role in its pathogenesis.

CASE REPORT

A boy was born at 38 weeks of gestation and presented with lethargy and hyporeactivity. No abnormalities were detected on prenatal ultrasonography. A loud systolic murmur was found. Bilateral femoral pulses were weak but palpable. Laboratory findings and a chest x-ray film revealed no abnormalities. Echocardiography showed normal intracardiac structures and some hypertrophy of the left ventricle. The aortic arch seemed intact, with normal anatomic origins of its main branches, but revealed an irregular adjacent mass in the convex part of the arch (Figure 1). The duct was wide open. High-resolution computed tomography confirmed the obstruction in the aortic arch and showed extension of the mass toward the medial lumen of the innominate artery with a critical ostial stenosis of the left carotid artery. Cerebral ultrasonography showed increased diastolic flow in the anterior pericallosal artery and hyporeflective swelling of the basal ganglia, indicating proximal stenosis with poststenotic dilatation and hypoxic edema, respectively. Therefore, urgent surgical exploration was performed. No external compression was found, and the aorta was opened. An irregular large fibrotic mass, adherent at the convex side of the aortic arch, obstructed the ostium of the brachiocephalic trunk and the left carotid artery completely (Figure 2). The ostium of the left subclavian artery was partially occluded. The mass was removed in toto. Postoperatively, the child could easily be weaned from inotropic support. Echocardiography showed good flow through the aortic arch and its main branches. Unfortunately, subsequent cerebral ultrasonography showed progressive, extensive brain damage. Magnetic resonance imaging (MRI) confirmed diffuse lesions of the white matter, with severe ischemic cortical necrosis in both hemispheres. Therapy was stopped after careful consultation with the parents. The child died on day 9. The pathologic features of the mass revealed thrombus, partially organized with central necrosis and peripheral calcifications. No autopsy was performed because the parents refused.

Thrombophilia screening test results (including antithrombin III, prothrombin, protein C, and protein S) were all within the normal range. No activated protein C resistance or anticardiolipine antibodies could be detected.

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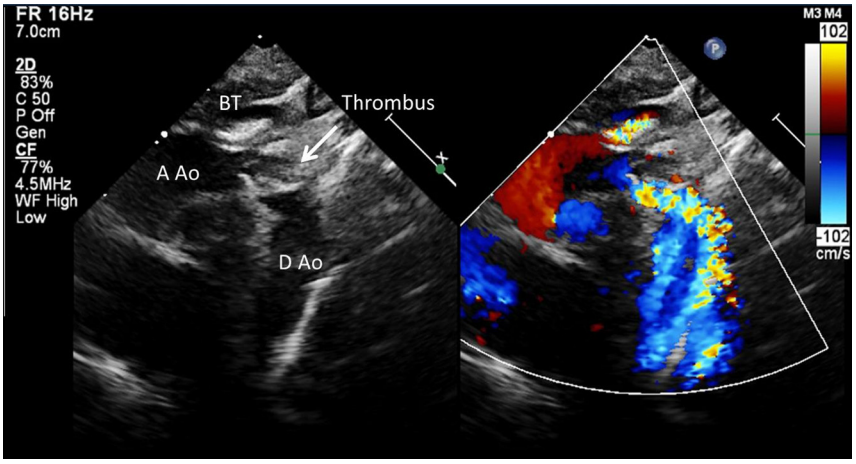

FIGURE 1. Preoperative echocardiography. *A Ao*, Ascending aorta; *BT*, brachiocephalic trunk; *D Ao*, descending aorta; *arrow*, thrombus located at the origin of the left carotid artery and left subclavian artery, without clear blood flow through these arteries. There was turbulent flow below the thrombus at the aortic cross level and turbulent flow at the brachiocephalic trunk (partially occluded).

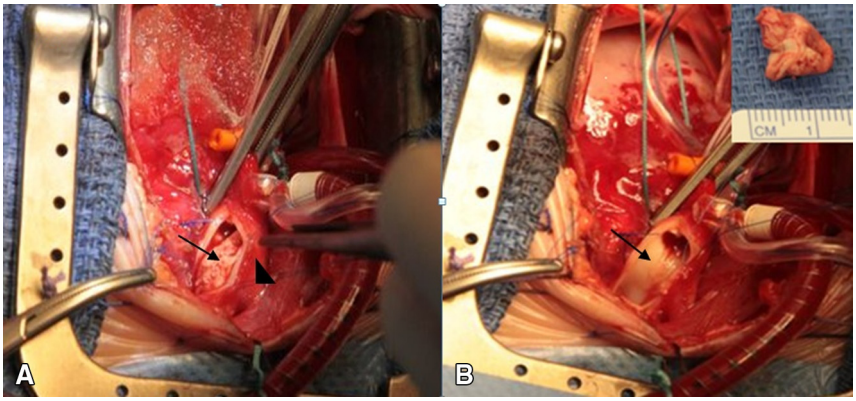

FIGURE 2. A, Perioperative image showing the thrombus in the aortic arch (*arrow*). *Arrowhead*, Brachiocephalic trunk. B, Aortic lumen after removal of the clot (*arrow*).

Screening results for lupus anticoagulant were negative. HCMV was detected in urine samples. The mother always tested HCMV seronegative.

DISCUSSION

Almost 90% of thromboses in neonates are associated with arterial or venous access devices. Other known risk factors in thromboembolic disease include maternal diabetes, sepsis, dehydration, polycythemia, poor cardiac output, and perinatal asphyxia.[1] Deficits in antithrombin or proteins C or S, the presence of factor V Leiden, and antiphospholipid syndrome have been previously described in neonates with thrombosis.[2] Almost all reports describe aortic thrombosis in the abdominal aorta. Thrombosis of the aortic arch has seldom been reported.

Our patient had no inherited thrombogenic risk factors. Urine cultures detected HCMV infection, which can be a possible trigger. Cerebral lesions of the white matter on MRI may be suggestive for HCMV infection. Cortical necrosis was too severe to differentiate between severe ischemia and ischemia with a contributive factor of HCMV infection. A link between aortic arch thrombosis in a neonate and HCMV infection in the absence of thrombogenic risk factors has previously been described only once.[2] HCMV can infect endothelial cells, which enhances thrombin generation. Alternatively, it is possible that the number of endothelial binding sites increases, thereby facilitating adhesion of inflammatory cells and platelets.

Treatment options include anticoagulation, thrombolytic therapy, and surgical thrombectomy.[3] The location and the characteristics of the thrombus seem to be important.[4,5] An old clot is less likely to resolve with fibrinolysis or anticoagulation because it is most often organized and calcified. Clots on a favorable location, with at least one carotid artery free from obstruction, have a more favorable outcome. Nevertheless, even when thrombectomy is successful, neurologic outcome may still be impaired because of cerebral ischemia or embolization. In our patient, we faced an old thrombus obstructing the complete aortic arch. The child did survive surgery but died because of neurologic impairment. In hindsight, it is not clear whether the neurologic deterioration could be attributed to preoperative cerebral hypoperfusion and embolization or whether this was the natural course of an active HCMV infection.

In conclusion, we report, to our knowledge, the second case of spontaneous aortic arch thrombosis associated with HCMV infection in a neonate. Consequently, HCMV detection should be included in routine laboratory testing whenever confronted with neonatal aortic arch thrombosis.

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